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Case Report

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Atypical arachnoid cyst with progression: a case report

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ABSTRACT

A 26-year-old female patient applied to the outpatient clinic in 2018 due to pain that had been present for a long time (16 years) and had recently spread from the top of the head to the nape of the neck. It was learned from the patient's anamnesis that her pain was relieved with analgesics. Although no pathological findings were found in the neurological examination, brain magnetic resonance imaging (MRI) was requested to reveal possible intracranial pathological conditions. In the MRI images taken, a cystic mass with a lobulated contour and thin septa of 24x31x40 mm and similar intensity to CSF was detected in the right parasagittal area posteriorly at the vertex level. The patient's control brain MRI images 3 months later showed no increase in cyst size. However, the patient's anamnesis, which came for a control in 2022, showed that his current complaints and headaches had increased. The MR images showed that the existing cyst had reached a size of approximately 60x35 mm. Medical treatment was initially planned for the patient (Acetazolamide, 750 mg/day). The MR taken one month later showed that there was no reduction in the cyst size and the optic discs were slightly swollen in the fundus examination. Considering the cyst growth, the patient's failure to benefit from medical treatment and the swelling noted in the optic discs, the patient was fitted with a cystoperitoneal shunt. The patient's early postoperative tomography image showed a significant reduction in the cyst (50.15x25.66) and a slightly swollen optic disc image with blurred borders was detected in the fundus examination. The patient's complaints regressed, and in the radiological images it was determined that the cyst continued to shrink [(45.8x23.19 mm) and (41.1x20.3 mm)], the shunt was in place, and she had no clinical complaints. As a result, it was argued that patients with atypically located arachnoid cysts, as in this case, should be taken under close clinical and radiological follow-up, and surgical treatment methods should definitely be considered in patients who are found to have an increase in cyst size and clinical findings during this follow-up and who do not respond despite medical treatment.

Keywords: Arachnoid cyst, cystoperitoneal shunt, papilledema

INTRODUCTION

Arachnoid cysts (ACs) are cystic dilatations that develop within the arachnoid membrane and are filled with cerebrospinal fluid (CSF).[1] Although it generally does not cause symptoms, it is often detected incidentally due to reasons such as increased access to health services and opportunities.[2,3] The most common symptom is headache. Although ACs generally do not change throughout life, examinations have demonstrated that some cysts grow. On the other hand, it has been reported that traumatic or spontaneous subdural hemorrhage may occur in patients followed with ACs.[1]

Arachnoid cysts generally do not require surgical treatment.[1,4] However, there is no clear procedure for surgical treatment to be applied to patients who are considered for surgery because of various reasons.[2,4-7]

In this case report, the clinic and treatment process of a patient with a growing arachnoid cyst are discussed with the literature.

CASE

A 26-year-old female patient applied to the outpatient clinic in 2018 due to pain that had been present for a long time (16 years) and had recently spread from the top of her head to her nape. It was learned from the patient's anamnesis that her pain was relieved with analgesics. Although no pathological findings were found in the neurological examination, brain

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MRI was requested to reveal possible intracranial pathological conditions. In the MR images, a cystic mass (arachnoid cyst?) measuring 24x31x40 mm (Figure 1A) with lobulated contours and thin septa, suppressed in FLAIR and of similar intensity to CSF, was observed in the right parasagittal area posteriorly at the vertex level. The patient was recommended to be followed up in 3 months. No additional pathological findings were found in the patient's examination in 2019, and no increase in cyst size was seen in the brain MRI images. Since no increase in cyst size was detected and there was no deterioration in the neurological level, surgical intervention was not considered, and follow-up was recommended. The patient was asked to come for a check-up after one year.

There were no new pathological findings in the patient's examination in 2021. In the brain MRI images, it was seen that the cystic mass persisted, and its dimensions were 34.50x 16.66 mm. The patient was recommended to follow up again.

It was learned from the anamnesis of the patient who came for a check-up in 2022 that his current complaints increased, her headache started from the nape of her neck and spread to the top of head and then hit her eyes and was occasionally triggered by light and sound, but she did not have complaints that increased with Valsalva, and she did not have any history of seizures, syncope or new trauma. The patient also stated that there was ringing in her right ear but no hearing problem. The patient's neurological examination revealed that there was no neurological deficit, her muscle strength was normal, and there were no pathological reflexes.

The patient underwent a control brain MRI for her current complaints. The MRI images showed that the existing cyst had reached approximately 60x35 mm in size. (Figure 1B). Medical treatment was initially planned for the patient and acetazolamide (750 mg/day) was started. The patient was advised to have visual field, visual acuity and fundus examination at the ophthalmology examination department. As a result of the examination and tests performed one month later, it was seen that the cyst size continued to grow, and the optic discs were slightly swollen in the fundus examination performed by the ophthalmologist (Figure 1C, Figure 1D). Considering the growth of the cyst, the patient's failure to benefit from medical treatment, and the swelling noted in the optic discs, surgical intervention was recommended to the patient.

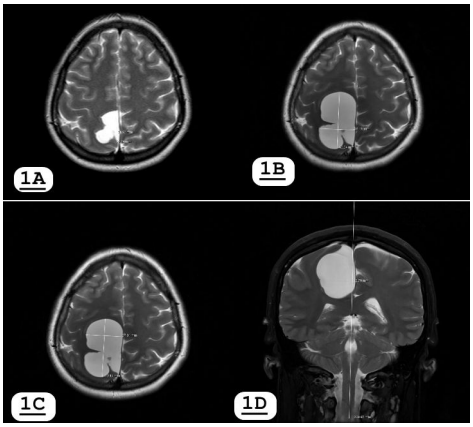

Figure 1. In the images, the arachnoid cyst image of the patient at the time of initial diagnosis (1A), the image with increased size during follow-up (1B), and the image taken after acetazolamide treatment (1C and 1D) are shown in T2-weighted MR images

The patient was placed on the operating table under general anesthesia in a supine position with the head slightly deviated to the left. Following the necessary field sterilization, the patient was covered in a sterile manner. The skin flap with the base remaining in the occipital was turned over in a C shape to the right Frazier point. One hole was opened with a high-speed drill to the Frazier point. A 3 cm skin incision was made from the right Mc Burney point. The peritoneum was seen by passing the subcutaneous layers. The peritoneal end of the shunt was advanced subcutaneously with the help of a shunt passer. The cranial end was sent to the cyst. Pressurized shunt flow was observed. The cranial end was connected to a medium-pressure dome. Subsequently, the peritoneal end was connected. Active shunt flow was observed. The catheter tip was sent into the abdomen through the previously opened peritoneal tunnel. The layers were closed in accordance with the procedure. Prophylactic antibiotic therapy was provided with 1 gram of ceftriaxone in the preoperative and postoperative processes.

No complications were observed in the patient who was hospitalized for 4 days after the operation. Postoperative tomography image of the patient revealed a significant reduction in the cyst (50.15x25.66) (Figure 2A). Fundus examination performed before discharge revealed a slightly swollen optic disc image with blurred borders. Postoperatively, the patient reported a significant reduction in headaches and improvement in blurred vision. In the patient's follow-ups, it was reported that the cyst continued to shrink (45.8x23.19 mm) and (41.1x20.3 mm) the shunt was in place, and that he had no clinical complaints (Figure 2B, Figure 2C).

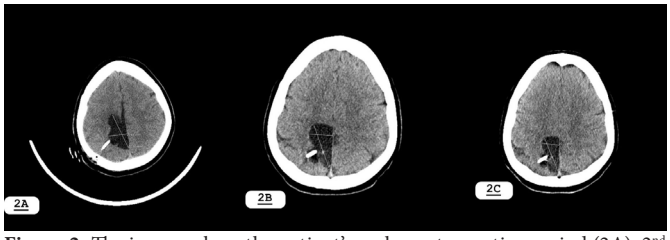

Figure 2. The images show the patient's early postoperative period (2A), 2nd month (2B) and 9th month (2C) CT images taken during clinical follow-up

DISCUSSION

Arachnoid cysts, described more than 150 years ago, are of two types: primary cysts and secondary cysts. Primary cysts occur congenitally after the division of the arachnoid membrane in the intrauterine period. Secondary cysts occur due to later factors such as trauma, infection, and surgery.[4,6] It constitutes approximately 1% of lesions causing intracranial mass and similar effects and generally occurs in the second decade of life.[8] The incidence is 1.4% [may be up to 2.6% in pediatric patients[9] and is more common in women.[6]

During pregnancy, after the 15th week of the fetus, the subarachnoid membrane is formed and then CSF is secreted. The secreted CSF contributes to the development of the network structure in the subarachnoid space and also helps the development of the arachnoid. First trapped in the ventricles, CSF later replaces the extracellular ground substance and begins to circulate freely in the subarachnoid space. As the arachnoid membrane develops, it can be trapped inside the contractions that occur. This cystic pocket is separated from the arachnoid membrane by the absence of trabecules crossing the cyst without any pathological changes in the underlying pia and cortex. All these conditions can

express the formation of ACs.[1,4,10] Briefly, the formation of this cyst may be caused by abnormal division or abnormal release of CSF.[1]

Arachnoid cysts can be located in different parts of the brain. The middle cranial fossa (MCF) is the most common location with 50% and is more common in men.[1,3,4,6,8-10] There are 3 types of cysts in MCF according to Galassi classification. Type-1 cysts are found in the anterior middle cranial fossa (MCF-A) and are asymptomatic. Type-2 cysts extend above the Sylvian fissure and are usually seen on the left. In some cases, it is seen that they can change the location of the temporal lobe. Type-3 cysts completely occupy the MCF and affect the parietal and frontal lobes along with the temporal lobe.[2,4,6] The frequency of ACs in the posterior cranial fossa (PCF) is 38%. However, ACs can also be seen in the quadrigeminal cistern, retro cerebellar area, cerebellopontine angle, and suprasellar areas, and the frequency of occurrence in these areas is approximately 10%. In some areas such as the interhemispheric fissure, ventricles, and cerebral convexities, the frequency of ACs is less.[1,3,4,6,8-10] Interhemispheric cysts were examined by Mori et al.[6] in two classes as parasagittal and midline cysts. Parasagittal cysts are unilateral; they generally do not show hydrocephalus because they are not close to the ventricle. Midline cysts are complex, multiloculated cysts. They occur with hydrocephalus. They generally show close proximity to the cyst at the level of the roof of the third ventricle. Midline cysts, unlike parasagittal cysts, may be associated with agenesis of the corpus callosum.

Many ACs do not change throughout life. However, studies have shown that some cysts grow. There are three possible reasons for growth: (a) a ball mechanism that allows CSF to enter the cyst, (b) abnormal production of CSF within the cyst, and (c) an osmotic gradient difference resulting from a high protein content within the cyst.[1,2,7,8] On the other hand, the gradient difference proposal is currently less acceptable. The reason for this is that the cyst content cannot be distinguished biochemically from CSF, the amount evacuated by shunt is generally equal to the daily CSF production, and there are publications stating that it has lost its validity and that the ball-valve mechanism is the main factor in cyst growth.[2,7] ACs usually do not cause symptoms, so they are diagnosed incidentally. In fact, they are often detected after head trauma in adults.[8,10] On the other hand, when the symptoms are examined from pediatric age group to adult, there is a wide range such as headache (66%), dizziness, nausea, vomiting, macrocephaly, hydrocephalus, papilledema, seizures, mental status changes.[2,3,6,8]

ACs are generally followed periodically, and conservative approaches are at the forefront. However, surgeons agree that patients with neural compression, persistent seizures, mass effect, focal neurologic deficits, and signs and symptoms of intracranial hypertension should be operated on.[8,10] On the other hand, researchers think that the area where the ACs are located plays an important role in the surgical treatment to be chosen and outcome of operation.[2,6] There are multiple options for surgical treatment, including craniotomy, open cyst fenestration, stereotactic cyst aspiration, endoscopic cyst fenestration, and shunt application. There is no consensus on which of these options is superior.[5,9] The main purpose of treatment is to eliminate the pressure created by the accumulated CSF and the symptoms related to it. There are publications recommending endoscopic fenestration in ACs in the suprasellar and quadrigeminal areas. There are also publications that consider endoscopic procedures to be more beneficial in terms of avoiding shunt and craniotomy complications.[7] The best results in terms of endoscopy have been reported in suprasellar cysts. Endoscopy is controversial in patients with middle fossa cysts; some authors prefer microsurgery over endoscopic surgery for cysts in this location.[6] There are studies that prioritize shunt application because of its low recurrence and complication rate, its effect on cyst reduction, and its ability to keep hydrocephalus and intracranial hypertension under control.[3,5,10] In addition, the fact that it is a relatively simple technique and that intraoperative guidance (ultrasonography and neuronavigation) can be used also makes shunt use advantageous.[8] It has also been stated that the superficiality of the cyst is an important factor in shunt preference.[2] In another study, it has been stated that shunt preference should be the first choice in elderly patients and complications are less common.[3]

Our patient was 26-years-old when diagnosed in the outpatient clinic and her gender was female. The location and structure of the cyst (frontoparietal area, multilobule) were atypical when compared to the literature. Although many arachnoid cysts are generally asymptomatic, the main complaint of the patient was headache. However, considering that the patient re-applied to the outpatient clinic with an increase in headaches, it should be considered that the patient's complaints may change during the follow-up period. During this time, the growth of the cyst observed in the radiological imaging and the increase in the patient's clinical complaints in direct proportion to this should be taken into account when evaluating the patient. However, the lack of response to the given medical treatment, failure to reduce in the cyst borders despite the medical treatment and the continuing complaints led to the consideration of the surgical option. The cyst in the patient was atypical, multiloculated, rare and growth faster which distinguishes it from other ACs. In addition, the presence of an atypical cyst is also important because it affects the clinical course of the disease, its progression and the surgical treatment to be chosen.

CONCLUSION

Although ACs are generally kept under clinical follow-up because they are asymptomatic, surgery may be performed if they grow and if signs related to increased intracranial pressure occur. The surgical method to be applied is determined by the location of the cyst and the surgeon's preference. In the case report mentioned in this article, cyst growth, increasing headache and papilledema occurred; in the surgical option, cystoperitoneal shunt was preferred and successfully applied due to the low probability of ACs recurrence, not being a high-level invasive procedure, tolerable complications and the location of the cyst which is atypical and close to the surface. Although it has been stated in previous publications that it is the first choice in the elderly patient population, its successful use in a superficial cyst in a young patient shows that the surgical option chosen for the patient is correct. This situation reveals the importance of the localization of the cyst and the surrounding anatomical structures in the treatment option. In postoperative observations, it was observed that the patient's complaints decreased, and the cyst shrank and the patient did not develop any complications.

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